

Amendments to the Claims:

In the claims:

Claim 1 (Previously Presented): A transgenic mouse wherein both alleles encoding Alpha Hemoglobin Stabilizing Protein (AHSP) have been disrupted via targeted insertion of a transgene wherein said mouse does not express a functional mouse Alpha Hemoglobin Stabilizing Protein (AHSP) protein and erythrocytes obtained from said mouse exhibit one or more characteristics selected from the group consisting of abnormal spiculated morphology, reduced lifespan, increased production of reactive oxygen species (ROS), and precipitated hemoglobin.

Claim 2 (Previously Presented): The transgenic mouse of claim 1, wherein said mouse transmits said transgene to its offspring.

Claim 3 (Previously Presented): The transgenic mouse of claim 1, wherein said transgene has been introduced into an ancestor of said mouse at an embryonic stage following microinjection of embryonic stem cells into a mouse blastocyst.

Claim 4 (Previously Presented): The transgenic mouse of claim 1, wherein said transgene has been introduced into an ancestor of said mouse at an embryonic stage following co-incubation of embryonic stem cells with a fertilized egg or morula.

Claim 5 (Previously Presented): A transgenic mouse wherein one allele of its endogenous Alpha Hemoglobin Stabilizing

Protein (AHSP) gene has been disrupted via targeted insertion of a transgene wherein said mouse exhibits AHSP haploinsufficiency and has an elevated reticulocyte count.

Claim 6 (Previously Presented): The transgenic mouse of claim 5, wherein said mouse and transmits said transgene to its offspring.

Claim 7 (Previously Presented): The transgenic mouse of claim 5, wherein said transgene has been introduced into an ancestor of said mouse at an embryonic stage following microinjection of embryonic stem cells into a mouse blastocyst.

Claim 8 (Previously Presented): The transgenic mouse of claim 5, wherein said transgene has been introduced into an ancestor of said mouse at an embryonic stage following co-incubation of embryonic stem cells with a fertilized egg or morula.

Claim 9 (Currently Amended): A method for screening for therapeutic agents which affect Alpha Hemoglobin Stabilizing Protein (AHSP) ~~activity~~, deficiency-related phenotypes comprising:

a) administering a test compound to the transgenic mouse of claim 1;

b) assessing said mouse for an alteration in an Alpha Hemoglobin Stabilizing Protein (AHSP) deficiency phenotype ~~activity~~.

Claim 10 (Original): The method of claim 9, wherein said activity is selected from the group consisting of α -hemoglobin binding, and α -hemoglobin synthesis.

Claim 11 (Currently Amended): A method for assessing the activity of a compound useful for the treatment and/or prevention of an AHSP related disorder, comprising:

- a) providing at least one transgenic mouse as claimed in claim 1;
- b) administering a test compound to said mouse; and
- ~~c)~~ c) assessing the erythrocytes of said mouse for an alteration in one or more characteristics selected from the group consisting of abnormal spiculated morphology, reduced lifespan, increased production of reactive oxygen species (ROS), and precipitated hemoglobin, thereby identifying agents useful for treatment of an AHSP related disorder.

Claim 12 (Original): The method of claim 11, said method further comprising administration of said test compound to a control mouse to assess toxicity of said test compound.

Claims 13-38 (Cancelled)

Claim 39 (Currently amended): The method of claim 9, wherein said test compound alters one or more AHSP deficiency characteristics selected from the group consisting of abnormal speculated morphology, reduced erythrocyte lifespan, increased production of reactive oxygen species (ROS), and precipitated hemoglobin in the erythrocytes of said mice.